General

Guideline Title

Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review). Report of the Quality Standards Subcommittee of the American Academy of Neurology.

Bibliographic Source(s)

Miller RG, Jackson CE, Kasarskis EJ, England JD, Forshew D, Johnston W, Kalra S, Katz JS, Mitsumoto H, Rosenfeld J, Shoesmith C, Strong MJ, Woolley SC, Quality Standards Subcommittee of the American Academy of Neurology. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology. 2009 Oct 13;73(15):1218-26. [40 references] PubMed

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Miller RG, Rosenberg JA, Gelinas DF, Mitsumoto H, Newman D, Sufit R, Borasio GD, Bradley WG, Bromberg MB, Brooks BR, Kasarskis EJ, Munsat TL, Oppenheimer EA. Practice parameter: the care of the patient with amyotrophic lateral sclerosis (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology: ALS Practice Parameters Task Force. Neurology 1999 Apr 22;52(7):1311-23. [112 references]

The American Academy of Neurology reaffirmed the currency of this guideline in 2013.

Recommendations

Major Recommendations

Definitions of the levels of the recommendations (A, B, C, U) and classification of the evidence (Class I-IV) are provided at the end of the "Major Recommendations" field.

Slowing the Disease Process

What is the effect of riluzole on slowing the disease process or prolonging survival in amyotrophic lateral sclerosis (ALS)?

Conclusion

Riluzole is safe and effective for slowing disease progression to a modest degree in ALS (4 Class I studies).

Recommendation

Riluzole should be offered to slow disease progression in patients with ALS (Level A).

Does lithium carbonate prolong survival or slow disease progression in ALS?

Conclusions

There are inadequate data on the effectiveness of lithium carbonate (1 Class III study).

Recommendations

There are insufficient data at this time to support or refute treatment with lithium carbonate in patients with ALS (Level U).

Nutrition

What is the effect of enteral nutrition administered via percutaneous endoscopic gastrostomy (PEG) on weight stability?

Conclusion

Enteral nutrition administered via PEG is probably effective in stabilizing body weight/body mass index (2 Class II, 7 Class III studies).

Recommendation

In patients with ALS with impaired oral food intake, enteral nutrition via PEG should be considered to stabilize body weight (Level B).

When is PEG indicated in ALS?

Conclusions

There are no studies of ALS-specific indications for the timing of PEG insertion, although patients with dysphagia will possibly be exposed to less risk if PEG is placed when forced vital capacity (FVC) is above 50% of predicted (1 Class III study).

Recommendation

There are insufficient data to support or refute specific timing of PEG insertion in patients with ALS (Level U).

What is the efficacy of nutritional support via PEG in prolonging survival?

Conclusions

Studies using appropriate controls or multivariate analysis demonstrated that PEG is probably effective in prolonging survival in ALS, although insufficient data exist to quantitate the survival advantage (2 Class II studies).

Recommendation

PEG should be considered for prolonging survival in patients with ALS (Level B).

What is the effect of enteral nutrition delivered via PEG on quality of life?

Conclusion

No evidence exists regarding the effect of enteral nutrition on quality of life.

Recommendations

There are insufficient data to support or refute PEG for improving quality of life in patients with ALS (Level U).

What is the efficacy of vitamin and nutritional supplements on prolonging survival or quality of life?

Conclusions

- Creatine, in doses of 5–10 g daily, is established as ineffective in slowing the rate of progression or in improving survival in ALS (2 Class I studies).
- Vitamin E 5,000 mg/day plus riluzole is probably ineffective in improving survival or functional outcomes (1 Class I study). Vitamin E (1,000 mg/day plus riluzole) was marginally effective in slowing the progression of ALS from milder to more severe ALS health states using a single measure but is ineffective using multiple other measures (1 Class I study).

Recommendations

Creatine, in doses of 5–10 g daily, should not be given as treatment for ALS because it is not effective in slowing disease progression (Level A). High-dose vitamin E should not be considered as treatment for ALS (Level B), while the equivocal evidence regarding low-dose vitamin E permits no recommendation (Level U).

Respiratory Management

What are the optimal pulmonary tests to detect respiratory insufficiency?

Conclusions

- Nocturnal oximetry and maximal inspiratory pressure (MIP) are possibly more effective in detecting early respiratory insufficiency than erect FVC (2 Class III studies).
- Supine FVC is possibly more effective than erect FVC in detecting diaphragm weakness and correlates better with symptoms of nocturnal hypoventilation (2 Class III studies).
- Sniff transdiaphragmatic pressure (Pdi) and sniff nasal pressure (SNP) are possibly effective in detecting hypercapnia and nocturnal hypoxemia (2 Class III studies).

Recommendations

- Nocturnal oximetry may be considered to detect hypoventilation (regardless of the FVC) (Level C).
- Supine FVC and MIP may be considered useful in routine respiratory monitoring, in addition to the erect FVC (Level C).
- SNP may be considered to detect hypercapnia and nocturnal hypoxemia (Level C).

Does noninvasive ventilation (NIV) improve respiratory function or increase survival?

Conclusion

NIV is probably effective in prolonging survival (1 Class I, 3 Class III studies) and in slowing the rate of FVC decline (1 Class I, 1 Class III study).

Recommendations

- NIV should be considered to treat respiratory insufficiency in ALS, both to lengthen survival (Level B) and to slow the decline of FVC (Level B).
- NIV may be considered to improve quality of life (Level C).

How do invasive and noninvasive ventilation affect quality of life?

Conclusions

- NIV is possibly effective in raising quality of life (QOL) for patients with ALS who have respiratory insufficiency (5 Class III studies).
- Tracheostomy invasive ventilation (TIV) is possibly effective in preserving QOL for patients with ALS, but possibly with a greater burden for their caregivers (2 Class III studies).

Recommendations

- NIV may be considered to enhance QOL in patients with ALS who have respiratory insufficiency (Level C).
- TIV may be considered to preserve QOL in patients with ALS who want long-term ventilatory support (Level C).

What factors influence acceptance of invasive and noninvasive ventilation?

Conclusions

- Nocturnal oximetry is possibly effective in detecting early respiratory insufficiency and the early use of NIV possibly increases compliance (2 Class III studies).
- Bulbar involvement and executive dysfunction possibly lower compliance with NIV (2 Class III studies).

Recommendation

NIV may be considered at the earliest sign of nocturnal hypoventilation or respiratory insufficiency in order to improve compliance with NIV in patients with ALS (Level C).

What is the efficacy of targeted respiratory interventions for clearing secretions?

Conclusions

- Mechanical insufflation/exsufflation (MIE) is possibly effective for clearing upper airway secretions in patients with ALS who have reduced peak cough flow, although the clinically meaningful difference is unknown (4 Class III studies).
- High frequency chest wall oscillation (HFCWO) is unproven for adjunctive airway secretion management (2 Class III studies with conflicting results).

Recommendations

- MIE may be considered to clear secretions in patients with ALS who have reduced peak cough flow, particularly during an acute chest infection (Level C).
- There are insufficient data to support or refute HFCWO for clearing airway secretions in patients with ALS (Level U).

Definitions:

Classification of Evidence for Studies of Therapeutic Intervention

Class I = A randomized, controlled clinical trial of the intervention of interest with masked or objective outcome assessment, in a representative population. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences. The following are also required:

a. Concealed allocation
b. Primary outcome(s) clearly defined
c. Exclusion/inclusion criteria clearly defined
d. Adequate accounting for drop-outs (with at least 80% of enrolled subjects completing the study) and cross-overs with numbers sufficiently low to have minimal potential for bias
e. For non-inferiority or equivalence trials claiming to prove efficacy for one or both drugs, the following are also required*:
    1. The authors explicitly state the clinically meaningful difference to be excluded by defining the threshold for equivalence or non-inferiority.
    2. The standard treatment used in the study is substantially similar to that used in previous studies establishing efficacy of the standard treatment. (e.g., for a drug, the mode of administration, dose and dosage adjustments are similar to those previously shown to be effective).
    3. The inclusion and exclusion criteria for patient selection and the outcomes of patients on the standard treatment are comparable to those of previous studies establishing efficacy of the standard treatment.
    4. The interpretation of the results of the study is based upon a per protocol analysis that takes into account dropouts or crossovers.

Class II = A randomized controlled clinical trial of the intervention of interest in a representative population with masked or objective outcome assessment that lacks one criteria a–e above or a prospective matched cohort study with masked or objective outcome assessment in a representative population that meets b–e above. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences.

Class III = All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome is independently assessed, or independently derived by objective outcome measurement.**

Class IV = Studies not meeting Class I, II, or III criteria including consensus or expert opinion.

*Note that numbers 1-3 in Class Ie are required for Class II in equivalence trials. If any one of the three is missing, the class is automatically downgraded to Class III.

**Objective outcome measurement: an outcome measure that is unlikely to be affected by an observer's (patient, treating physician, investigator) expectation or bias (e.g., blood tests, administrative outcome data).

Classification of Evidence for Diagnostic Accuracy

Class I = A cohort study with prospective data collection of a broad spectrum of persons with the suspected condition, using an acceptable reference standard for case definition. The diagnostic test is objective or performed and interpreted without knowledge of the patient's clinical status. Study results allow calculation of measures of diagnostic accuracy.

Class II = A case control study of a broad spectrum of persons with the condition established by an acceptable reference standard compared to a broad spectrum of controls or a cohort study where a broad spectrum of persons with the suspected condition where the data was collected

retrospectively. The diagnostic test is objective or performed and interpreted without knowledge of the disease status. Study results allow calculation of measures of diagnostic accuracy.

Class III = A case control study or a cohort study where either the persons with the condition or the controls are of a narrow spectrum. The condition is established by an acceptable reference standard. The reference standard and diagnostic test are objective or performed and interpreted by different observers. Study results allow calculation of measures of a diagnostic accuracy.

Class IV = Studies not meeting Class I, II, or III criteria including consensus, expert opinion or a case report.

Classification of Recommendations

Level A = Established as effective, ineffective, or harmful (or established as useful/predictive or not useful/predictive) for the given condition in the specified population. (Level A rating requires at least two consistent Class I studies.*)

Level B = Probably effective, ineffective, or harmful (or probably useful/predictive or not useful/predictive) for the given condition in the specified population. (Level B rating requires at least one Class I study or two consistent Class II studies.)

Level C = Possibly effective, ineffective, or harmful (or possibly useful/predictive or not useful/predictive) for the given condition in the specified population. (Level C rating requires at least one Class II study or two consistent Class III studies.)

Level U = Data inadequate or conflicting; given current knowledge, treatment (test, predictor) is unproven.

*In exceptional cases, one convincing Class I study may suffice for an "A" recommendation if: 1) all criteria are met, 2) the magnitude of effect is large (relative rate improved outcome >5 and the lower limit of the confidence interval is >2).

Clinical Algorithm(s)

The original guideline document contains clinical algorithms for:

- Nutrition management
- Respiratory management

Scope

Disease/Condition(s)

Amyotrophic lateral sclerosis (ALS; also known as Lou Gehrig's disease)

Guideline Category

Assessment of Therapeutic Effectiveness

Counseling

Evaluation

Management

Risk Assessment

Treatment

Clinical Specialty

Family Practice

Internal Medicine

Neurology

Nutrition

Pulmonary Medicine

Intended Users

Advanced Practice Nurses

Dietitians

Pharmacists

Physician Assistants

Physicians

Respiratory Care Practitioners

Guideline Objective(s)

To systematically review evidence bearing on the management of patients with amyotrophic lateral sclerosis (ALS)

Target Population

Patients with amyotrophic lateral sclerosis

Interventions and Practices Considered

1. Pharmacotherapy
    - Riluzole
    - Lithium carbonate (no recommendation made)
2. Nutrition
    - Percutaneous endoscopic gastrostomy (PEG)
    - Timing of PEG (no recommendation made)
    - High-dose vitamins, minerals, and other nutriceuticals
        - Creatine (not recommended)
        - Vitamin E (no recommendation made)
3. Respiratory management
    - Optimal pulmonary function tests
        - Forced vital capacity (FVC)
        - Maximal inspiratory pressure (MIP)
        - Nocturnal oximetry
        - Sniff transdiaphragmatic pressure (sniff Pdi)
        - Sniff nasal pressure
        - Bicarbonate and serum chloride levels
        - Peak cough expiratory flow (PCEF)
    - Respiratory assistance
        - Noninvasive ventilation (NIV)
        - Tracheostomy invasive ventilation (TIV)
        - Supplemental oxygen
        - Ventilatory compliance
    - Clearing secretions (targeted respiratory intervention)

- Mechanical insufflation/exsufflation
- High frequency chest wall oscillation
- Medications with mucolytics
- Anticholinergic bronchodilator

Major Outcomes Considered

- Weight (stability, gain)
- Quality of life
- Degree of symptom (pain, anxiety, dyspnea) control
- Rate of disease progression
- Changes in pulmonary function tests
- Respiratory failure
- Compliance with noninvasive ventilation
- Patient survival

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

2009 Guideline

The authors searched OVID, MEDLINE EMBASE, CINAHL, Science Citation Index, BIOETHICSLINE, International Pharmaceutical Abstracts (IPAB), OVID Current contents, Medline-ProQuest, EIFL, and INVEST from 1998 through September 2007, combining the words ALS, Lou Gehrig's disease, and motor neuron disease with the following words using AND: respiratory, respiratory failure, respiratory insufficiency, nutrition, enteral nutrition, malnutrition, weight loss, gastrostomy, clinical trials, mechanical insufflation-exsufflation, high frequency chest wall oscillation, Vest, Bipap, tracheostomy ventilation, dysphagia, mechanical ventilation, noninvasive ventilation, hypoventilation, bronchial secretions, sleep-disordered breathing, and breath stacking. They reviewed the abstracts of these articles and examined 142 articles in their entirety.

2013 Reaffirmation

The guideline developer searched OVID MEDLINE, EMBASE, CINAHLScience Citation Index, BIOETHICSLINE, International Pharmaceutical Abstractions (IPAB), OVID Current contents, Medline-ProQuest, EIFL, and INVEST for studies published between 2010 and 2013 using the following search terms: ALS, Lou Gehrig's disease, and motor neuron disease with the following words using AND: respiratory, respiratory failure, respiratory insufficiency, nutrition, enteral nutrition, malnutrition, weight loss, gastrostomy, clinical trials, mechanical insufflation-exsufflation, high frequency chest wall oscillation, Vest, Bipap, tracheostomy, ventilation, dysphagia, mechanical ventilation, noninvasive ventilation, hypoventilation, bronchial secretions, sleep-disordered breathing, and breath stacking.

Number of Source Documents

Not stated

Methods Used to Assess the Quality and Strength of the Evidence

Expert Consensus

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Classification of Evidence for Studies of Therapeutic Intervention

Class I = A randomized, controlled clinical trial of the intervention of interest with masked or objective outcome assessment, in a representative population. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences. The following are also required:

a. Concealed allocation
b. Primary outcome(s) clearly defined
c. Exclusion/inclusion criteria clearly defined
d. Adequate accounting for drop-outs (with at least 80% of enrolled subjects completing the study) and cross-overs with numbers sufficiently low to have minimal potential for bias
e. For non-inferiority or equivalence trials claiming to prove efficacy for one or both drugs, the following are also required*:
    1. The authors explicitly state the clinically meaningful difference to be excluded by defining the threshold for equivalence or non-inferiority.
    2. The standard treatment used in the study is substantially similar to that used in previous studies establishing efficacy of the standard treatment. (e.g., for a drug, the mode of administration, dose and dosage adjustments are similar to those previously shown to be effective).
    3. The inclusion and exclusion criteria for patient selection and the outcomes of patients on the standard treatment are comparable to those of previous studies establishing efficacy of the standard treatment.
    4. The interpretation of the results of the study is based upon a per protocol analysis that takes into account dropouts or crossovers.

Class II = A randomized controlled clinical trial of the intervention of interest in a representative population with masked or objective outcome assessment that lacks one criteria a–e above or a prospective matched cohort study with masked or objective outcome assessment in a representative population that meets b–e above. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences.

Class III = All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome is independently assessed, or independently derived by objective outcome measurement.**

Class IV = Studies not meeting Class I, II, or III criteria including consensus or expert opinion.

*Note that numbers 1-3 in Class Ie are required for Class II in equivalence trials. If any one of the three is missing, the class is automatically downgraded to Class III.

**Objective outcome measurement: an outcome measure that is unlikely to be affected by an observer's (patient, treating physician, investigator) expectation or bias (e.g., blood tests, administrative outcome data).

Classification of Evidence for Diagnostic Accuracy

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Class II = A case control study of a broad spectrum of persons with the condition established by an acceptable reference standard compared to a broad spectrum of controls or a cohort study where a broad spectrum of persons with the suspected condition where the data was collected retrospectively. The diagnostic test is objective or performed and interpreted without knowledge of the disease status. Study results allow calculation of measures of diagnostic accuracy.

Class III = A case control study or a cohort study where either the persons with the condition or the controls are of a narrow spectrum. The condition is established by an acceptable reference standard. The reference standard and diagnostic test are objective or performed and interpreted by different observers. Study results allow calculation of measures of a diagnostic accuracy.

Class IV = Studies not meeting Class I, II, or III criteria including consensus, expert opinion or a case report.

Methods Used to Analyze the Evidence

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

The diagnostic and therapeutic classification schemes used to grade the articles are summarized in the "Rating Scheme for the Strength of the Evidence" field.

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

2009 Guideline

The American Academy of Neurology (AAN) assembled a panel of experts with expertise in amyotrophic lateral sclerosis (ALS). The Quality Standards Subcommittee of the American Academy of Neurology developed a set of clinical questions relevant to the evaluation of drug, nutritional, and respiratory management issues related to the care of patients with ALS. The strength of the practice recommendations was directly linked to the class of evidence using the scheme described in the "Rating Scheme for the Strength of the Recommendations" field.

2013 Reaffirmation

The AAN assesses their clinical practice guidelines every 2 years to determine whether new literature has been published that would warrant an update. The following steps are taken:

- Biennial correspondence is sent to all authors and the facilitator.
- An updated literature search and a review of methodological soundness are performed by a Guideline Development Subcommittee (GDS) member. (Note: The search should specifically seek to identify new evidence that would change the conclusions in the systematic review or recommendations in the CPG.)

All documents biennially reviewed by the GDS that don't require an update are reaffirmed. See the AAN Clinical Practice Guideline Process Manual for additional information.

Rating Scheme for the Strength of the Recommendations

Classification of Recommendations

Level A = Established as effective, ineffective, or harmful (or established as useful/predictive or not useful/predictive) for the given condition in the specified population. (Level A rating requires at least two consistent Class I studies.*)

Level B = Probably effective, ineffective, or harmful (or probably useful/predictive or not useful/predictive) for the given condition in the specified population. (Level B rating requires at least one Class I study or two consistent Class II studies.)

Level C = Possibly effective, ineffective, or harmful (or possibly useful/predictive or not useful/predictive) for the given condition in the specified population. (Level C rating requires at least one Class II study or two consistent Class III studies.)

Level U = Data inadequate or conflicting; given current knowledge, treatment (test, predictor) is unproven.

*In exceptional cases, one convincing Class I study may suffice for an "A" recommendation if: 1) all criteria are met, 2) the magnitude of effect is large (relative rate improved outcome >5 and the lower limit of the confidence interval is >2).

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

External Peer Review

Internal Peer Review

Description of Method of Guideline Validation

The guidelines were approved by the American Academy of Neurology (AAN) Quality Standards Subcommittee on November 5, 2008, by the Practice Committee on February 19, 2009, and by the Executive Board of the American Academy of Neurology on July 30, 2009.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see "Major Recommendations").

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

- Improved care and quality for life of people with amyotrophic lateral sclerosis
- Slowed disease progression
- Prolonged survival

Potential Harms

- Fatigue and nausea are known side effects of riluzole.
- Risks of percutaneous endoscopic gastrostomy (PEG) placement include laryngeal spasm, localized infection, gastric hemorrhage, failure to place PEG due to technical difficulties, and death due to respiratory arrest.

Qualifying Statements

Qualifying Statements

This statement is provided as an educational service of the American Academy of Neurology (AAN). It is based on an assessment of current scientific and clinical information. It is not intended to include all possible proper methods of care for a particular neurologic problem or all legitimate criteria for choosing to use a specific procedure. Neither is it intended to exclude any reasonable alternative methodologies. The American Academy of Neurology recognizes that specific patient care decisions are the prerogative of the patient and the physician caring for the patient, based on all of the circumstances involved. The clinical context section is made available in order to place the evidence-based guideline(s) into perspective with current practice habits and challenges. No formal practice recommendations should be inferred.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Clinical Algorithm

Patient Resources

Quick Reference Guides/Physician Guides

Resources

Slide Presentation

Staff Training/Competency Material

Wall Poster

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

End of Life Care

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

Miller RG, Jackson CE, Kasarskis EJ, England JD, Forshew D, Johnston W, Kalra S, Katz JS, Mitsumoto H, Rosenfeld J, Shoesmith C, Strong MJ, Woolley SC, Quality Standards Subcommittee of the American Academy of Neurology. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology. 2009 Oct 13;73(15):1218-26. [40 references] PubMed

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

1999 Apr (revised 2009 Oct 13; reaffirmed 2013 Nov)

Guideline Developer(s)

American Academy of Neurology - Medical Specialty Society

Source(s) of Funding

American Academy of Neurology (AAN)

Guideline Committee

Quality Standards Subcommittee

Composition of Group That Authored the Guideline

Guideline Authors: R.G. Miller, MD, FAAN; C.E. Jackson, MD, FAAN; E.J. Kasarskis, MD, PhD, FAAN; J.D. England, MD, FAAN; D. Forshew, RN; W. Johnston, MD; S. Kalra, MD; J.S. Katz, MD; H. Mitsumoto, MD, FAAN; J. Rosenfeld, MD, PhD, FAAN; C. Shoesmith, MD, BSc; M.J. Strong, MD; S.C. Woolley, PhD

Quality Standards Subcommittee Members 2007-2009: Jacqueline French, MD, FAAN (*Chair*); Charles E. Argoff, MD; Eric Ashman, MD; Stephen Ashwal, MD, FAAN (Ex-Officio); Christopher Bever, Jr., MD, MBA, FAAN; John D. England, MD, FAAN; Gary M. Franklin, MD, MPH, FAAN (Ex-Officio); Deborah Hirtz, MD, FAAN (Ex-Officio); Robert G. Holloway, MD, MPH, FAAN; Donald J. Iverson, MD, FAAN; Steven R. Messé, MD; Leslie A. Morrison, MD; Pushpa Narayanaswami, MD, MBBS; James C. Stevens, MD, FAAN (Ex-Officio); David J. Thurman, MD, MPH (Ex-Officio); Dean M. Wingerchuk, MD, MSc, FRCP(C); Theresa A. Zesiewicz, MD, FAAN

Financial Disclosures/Conflicts of Interest

Disclosure

Dr. Miller serves on the editorial board of the ALS Journal; received a speaker honorarium from the AANEM; served as a consultant to Celgene, Knopp Neurosciences Inc., Teva Pharmaceutical Industries Ltd., Taiji Biomedical, Inc., Sanofi-Aventis, Novartis, and Neuraltus; and receives research support from the NIH [R01 NS 44887 (PI)] and the Muscular Dystrophy Association (PI). Dr. Jackson serves as a consultant to Knopp Neurosciences Inc.; and receives research support from Knopp Neurosciences Inc., Insmed Inc., Solstice Neurosciences, Inc., the ALS Association, and the NIH NINDS [U01 NS042685-0 (Site PI), R01NS045087-01A2 (Site PI), and N01-AR-2250 (Site PI)]. Dr. Kasarskis serves as an Associate Editor for Amyotrophic Lateral Sclerosis; has received honoraria from the American Institute for Biological Studies (grant reviews); served as a consultant to Acceleron Pharma; holds equity in Amgen; and receives research support from the NIH/NINDS [R01-NS045087 (PI) and 1U01 NS049640 (Site PI). Dr. England serves as an Associate Editor for Current Treatment Options in Neurology; received a speaker honorarium from Teva Pharmaceutical Industries Ltd.; and serves as a consultant to Talecris. Ms. Forshew has served on a scientific advisory board for the ALS Association and receives research support from the Muscular Dystrophy Association. Dr. Johnston reports no disclosures. Dr. Kalra receives research support from the ALS Association of America and the ALS Society of Canada. Dr. Katz has received research support from Pfizer Inc. Dr. Mitsumoto served on scientific advisory boards for Avanir Pharmaceuticals, Knopp Neurosciences Inc., Neuralstem, Inc., Aisai Communication Technology Co., Ltd., and Otsuka Pharmaceutical Co., Ltd.; and receives research support from Avanir Pharmaceuticals, Teva Pharmaceutical Industries Ltd., Knopp Neurosciences Inc., Sanofi-Aventis, Athena Diagnostics, Inc., BioScrip, and the NIH/NINDS [DNA repository as a supplement and NIEHS Center grant]. Dr. Rosenfeld serves on the editorial board of Amyotrophic Lateral Sclerosis and has served as a consultant to Solstice Neurosciences, Inc. and Avicena Group, Inc. Dr. Shoesmith receives research support from the Muscular Dystrophy Association and her spouse is employed by Biovail Pharmaceuticals Canada. Dr. Strong serves on the editorial board of Amyotrophic Lateral Sclerosis. Dr. Woolley has received research support from Pfizer Inc., Eisai Inc., and the ALS Association (Co-I).

Conflict of Interest

The American Academy of Neurology is committed to producing independent, critical and truthful clinical practice guidelines (CPGs). Significant efforts are made to minimize the potential for conflicts of interest to influence the recommendations of this CPG. To the extent possible, the AAN

keeps separate those who have a financial stake in the success or failure of the products appraised in the CPGs and the developers of the guidelines. Conflict of interest forms were obtained from all authors and reviewed by an oversight committee prior to project initiation. AAN limits the participation of authors with substantial conflicts of interest. The AAN forbids commercial participation in, or funding of, guideline projects.

Drafts of the guideline have been reviewed by at least three AAN committees, a network of neurologists, Neurology® peer reviewers and representatives from related fields. The AAN Guideline Author Conflict of Interest Policy can be viewed at www.aan.com.

Guideline Endorser(s)

American Association of Neuromuscular and Electrodiagnostic Medicine - Medical Specialty Society

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Miller RG, Rosenberg JA, Gelinas DF, Mitsumoto H, Newman D, Sufit R, Borasio GD, Bradley WG, Bromberg MB, Brooks BR, Kasarskis EJ, Munsat TL, Oppenheimer EA. Practice parameter: the care of the patient with amyotrophic lateral sclerosis (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology: ALS Practice Parameters Task Force. Neurology 1999 Apr 22;52(7):1311-23. [112 references]

The American Academy of Neurology reaffirmed the currency of this guideline in 2013.

Guideline Availability

Electronic copies: A list of American Academy of Neurology (AAN) guidelines, along with a link to a Portable Document Format (PDF) file for this guideline, is available at the AAN Web site.

Print copies: Available from the AAN Member Services Center, (800) 879-1960, or from AAN, 201 Chicago Avenue South, Minneapolis, MN 55415.

Availability of Companion Documents

The following are available:

- Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review). AAN summary of evidence-based guideline for clinicians. St. Paul (MN): American Academy of Neurology. 2009 Apr. 2 p. Available in Portable Document Format (PDF) from the American Academy of Neurology (AAN) Web site.
- Practice parameter update: the care of the patient with amyotrophic lateral sclerosis. Case study. St. Paul (MN): American Academy of Neurology. 2009. 7 p. Available in PDF from the AAN Web site.
- Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review). Slide presentation. St. Paul (MN): American Academy of Neurology. 2009. 115 p. Available from the AAN Web site.
- Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review). Poster. St. Paul (MN): American Academy of Neurology. 2009. 1 p. Available in PDF from the AAN Web site.
- AAN guideline development process [online]. St. Paul (MN): American Academy of Neurology. Available from the AAN Web site.

Patient Resources

The following is available:

- Care of ALS: drug, symptom, nutritional, and breathing therapies. AAN summary of evidence-based guideline for patients and their families. St. Paul (MN): American Academy of Neurology (AAN). 2009. 2 p. Electronic copies: Available in Portable Document Format (PDF) from the AAN Web site .

Please note: This patient information is intended to provide health professionals with information to share with their patients to help them better understand their health and their diagnosed disorders. By providing access to this patient information, it is not the intention of NGC to provide specific medical advice for particular patients. Rather we urge patients and their representatives to review this material and then to consult with a licensed health professional for evaluation of treatment options suitable for them as well as for diagnosis and answers to their personal medical questions. This patient information has been derived and prepared from a guideline for health care professionals included on NGC by the authors or publishers of that original guideline. The patient information is not reviewed by NGC to establish whether or not it accurately reflects the original guideline's content.

NGC Status

This summary was completed by ECRI on February 12, 2002. The information was verified by the guideline developer as of March 29, 2002. This summary was updated by ECRI Institute on November 2, 2007, following the U.S. Food and Drug Administration advisory on Antidepressant drugs. This summary was updated by ECRI Institute on August 31, 2010. The currency of the guideline was reaffirmed by the developer in 2013 and this summary was updated by ECRI Institute on May 27, 2015.

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